

# CONGENITAL TRICUSPID ATRESIA WITH TRANSPOSITION OF THE GREAT VESSELS

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Congenital tricuspid atresia has been classified into two groups, those with and those without transposition of the great vessels (Kühne, 1906). Each of these groups is further subdivided according to whether there is an increase or a decrease in the pulmonary blood flow (Edwards and Burchell, 1949). Tricuspid atresia with transposition of the great vessels and an increase in pulmonary blood flow is uncommon and is associated with an average survival time of three months (Keith *et al.*, 1958). We have recently had the opportunity to study three hearts with this defect, one from a boy who survived for five years.

## CASE HISTORIES

*Case 1.* A female child aged 9 days was admitted to the Royal Maternity Hospital, Belfast in 1953. She was grossly cyanosed and dyspnoeic at rest, and had cardiac failure. Her condition deteriorated and she died one day after admission.

*Autopsy.* Apart from pneumonia in the upper lobe of the right lung the abnormal findings were confined to the heart and great vessels. There was tricuspid atresia, the remnants of the tricuspid valve being represented by a small depression in the floor of the right atrium. There was a large patent foramen ovale. The venous drainage to the atria was normal.

The mitral valve opening was large, being 4 cm. in circumference, and opened into a large ventricular cavity, the wall of which was 9 mm. thick. There was a large ventricular septal defect, the septum being represented by a small thick band of muscle, 7 mm. high, lying just below the great vessels. The great vessels were completely transposed. There was no pulmonary valve stenosis, the pulmonary trunk being almost twice the diameter of the aorta. There was a preductal coarctation through which a probe, 1 mm. in diameter, could just be passed. The ductus arteriosus was patent. The branches of the aorta were otherwise normal. The only histological feature of note was confirmation of the pneumonia in the lung. There were no pulmonary vascular lesions.

*Case 2.* A female child aged four days was admitted to the Ulster Hospital for Children and Women in 1958 with a history of cyanotic attacks while feeding. She was cyanosed and in congestive cardiac failure. Radiological examination of the chest showed generalized cardiac enlargement with much pulmonary plethora. In spite of therapy the child's condition worsened and she died at the age of 7 weeks.

*Autopsy.* The only abnormal findings were confined to the heart and great vessels. There was tricuspid atresia, a patent foramen ovale, and a wide mitral valve opening, 5 cm. in circumference. Both atria were dilated but had a normal venous drainage. The only evidence of ventricular separation was a thick band of muscle, 8 mm. high, just inferior to the origin of the great vessels. There was complete transposition of the great vessels. The aortic valve was 18 mm. and the pulmonary valve 24 mm. in circumference. No coarctation was present and the ductus arteriosus was closed. Histological examination of the lungs revealed no abnormal findings.

*Case 3.* A boy, aged five years and ten months, was admitted in 1958 to the Royal Belfast Hospital for Sick Children, having had a small hæmoptysis. His previous history consisted of two attacks of cardiac failure and cyanosis, the first at the age of six weeks and the second two months before admission, following an attack of whooping cough: both responded to treatment with digitalis. Exercise tolerance, though limited, was sufficient for ordinary activities, apart from the periods when the child was in cardiac failure.

On admission he was dyspnoëic at rest and cyanosed, with clubbing of the fingers. The venous pressure was raised, the liver was enlarged, and there were crepitations at both bases. There was marked cardiac enlargement. There was a rough systolic murmur at the lower end of the sternum, accompanied by a thrill, and an early diastolic murmur over the pulmonary area. Radiological examination of the chest showed generalized cardiac enlargement with considerable pulmonary plethora. An electrocardiogram recorded on admission showed the presence of sinus rhythm at a rate of 115 a minute and absence of the R waves over the right præcordium.

In spite of treatment with salt restriction, digitalis, and chlorothiazide, his condition rapidly deteriorated and he died two days after admission before further investigation was possible.

*Autopsy.* The relevant pathological findings were confined to the heart and lungs. The heart was enlarged and the main abnormalities are illustrated in Fig. 1 and 2. There was a normal venous return to both atria. There was tricuspid atresia. A large defect was present in the atrial septum in the region of the fossa ovalis due to gross fenestration of the septum primum (see in Fig. 1). The left atrium was

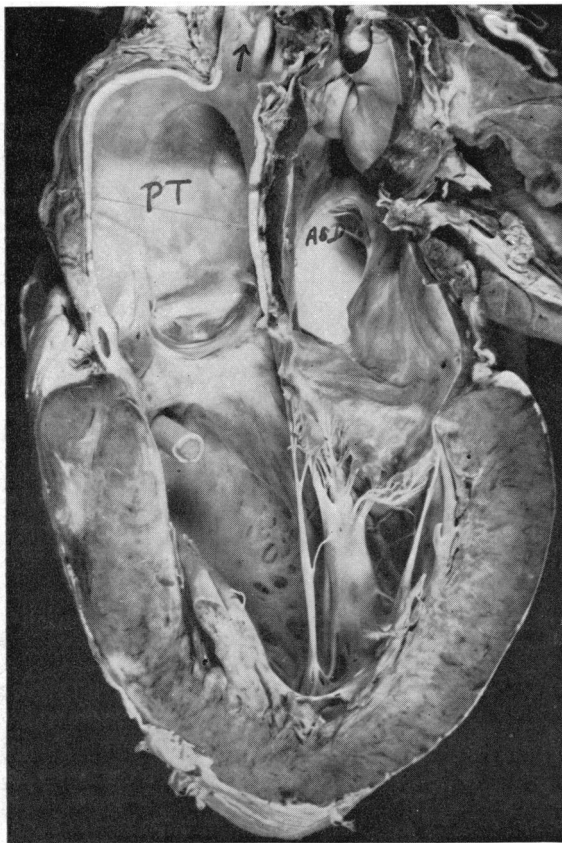


FIG. 1.—Photograph showing the left atrium, its septal defect (ASD), the mitral valve, and a large left ventricle. The large pulmonary trunk (PT) is arising from the left ventricle and a probe lies in the ventricular septal defect. The arrow indicates a patch of atheroma in the pulmonary artery. Case 3.

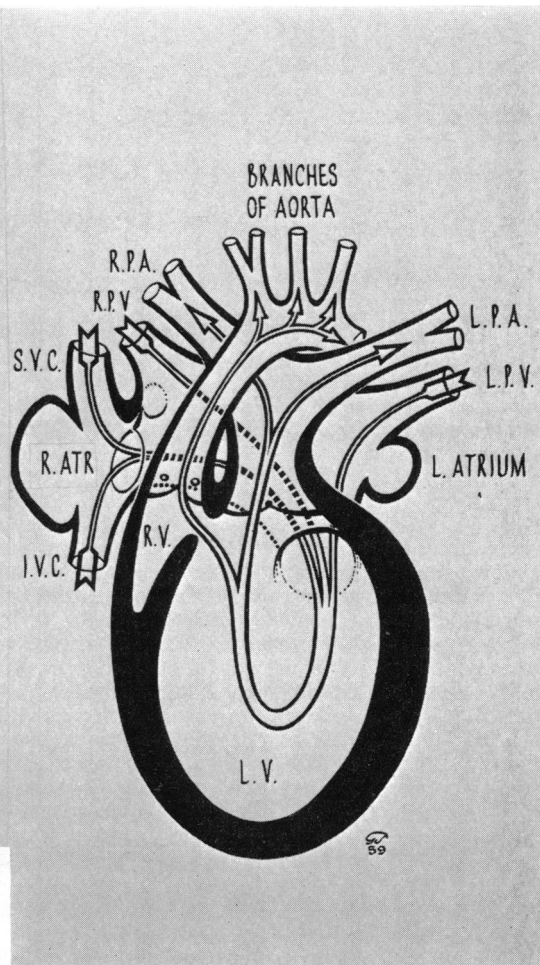


FIG. 2.—Diagram of the circulatory abnormalities in Case 3. The direction of blood flow is shown by the arrows. S.V.C. and I.V.C.=superior and inferior vena cava. R.P.A. and L.P.A.=right and left pulmonary artery. R.P.V. and L.P.V.=right and left pulmonary vein. R.V. and L.V.=right and left ventricle.

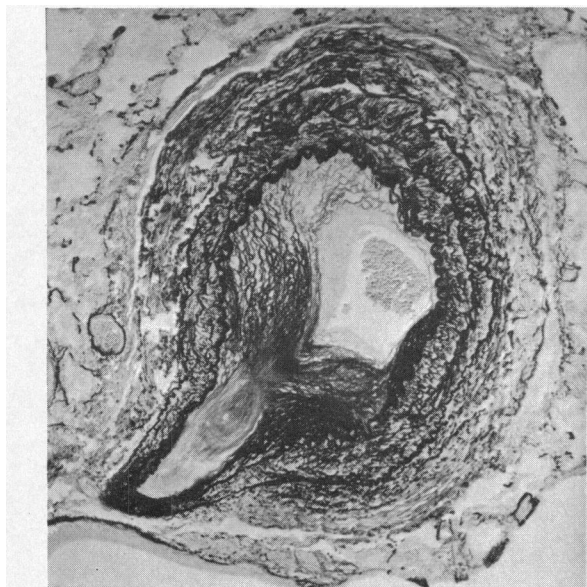


FIG. 3.—Case 3. Lung showing hypertrophy of the elastic and muscular tissue of the media of a medium-sized artery, with moderate elastic tissue proliferation of the intima. Hart van Geison:  $\times 75$ .

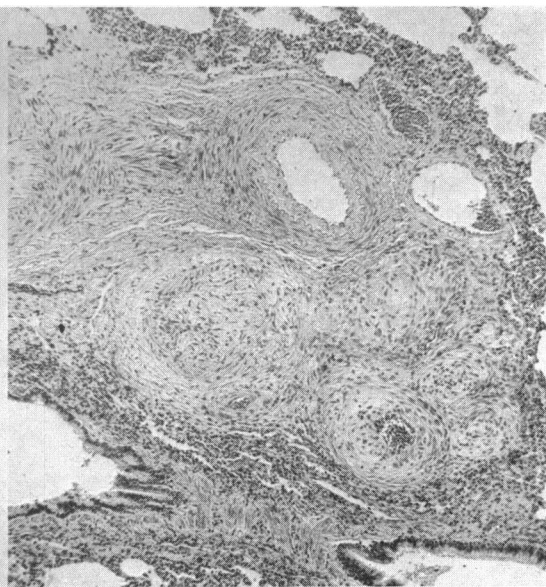


FIG. 4.—Case 3. Smaller arteries in the lungs, two of which are entirely occluded by fibrous tissue. Hæmatoxylin and eosin:  $\times 75$ .

moderately dilated and led into a wide mitral valve orifice which was 7 cm. in circumference. The left ventricle was dilated and hypertrophied (10–12 mm. thick). Most of the cardiac enlargement was due to the left ventricle.

There was complete transposition of the great vessels. The pulmonary trunk was large, being 6 cm. in circumference at the valve: it had a smooth intimal surface, but its main branches showed much atheroma (Fig. 1). There was a small oval ventricular septal defect, at the lower edge of the membranous septum but entirely bounded by muscle,  $4 \times 8$  mm. in size: it led into a small right ventricular cavity (Fig. 2). The endocardium surrounding the defect was pale and fibrosed. The aorta was relatively small, being 3.5 cm. in circumference at the valve. The ductus arteriosus was not patent and there was no coarctation. There was a small 8 mm. diameter healed infarct in the left kidney.

*Microscopical Examination.* All organs were examined microscopically and the only abnormal findings were found in the vascular system of the lungs. There was thickening of the intima due to atheroma in the large pulmonary arteries at the hilum of the lung. The walls of the medium-sized arteries showed moderate muscular hypertrophy and prominent elastic tissue hyperplasia in the media (Fig. 3). Many of the smaller arteries were occluded or narrowed by fibrous tissue which probably represented organized thrombi (Fig. 4). However, in a few of the medium-sized arteries this intimal thickening was partly composed of elastic tissue (Fig. 3). An occasional vessel showed well-marked arteritis with fibrin and polymorphonuclear leucocytes throughout the wall. Occasional giant cells were seen close to the walls of some of the arteries.

#### DISCUSSION

The three cases of tricuspid atresia presented are part of a collection of 175 autopsy specimens of congenital heart disease that were available for examination. Tricuspid atresia is a fairly uncommon form of congenital heart disease and is always, as in these cases, associated with an aperture in the atrial septum and a wide mitral valve opening. Each of our three cases had complete transposition of the great vessels and a wide pulmonary trunk.

We have been able to find 17 similar reported cases (Table I). These are described as tricuspid atresia type IIB by Edward and Burchell (1949).

As Keith *et al.* (1958) have stressed, few of this group lived for longer than three months (Table I). In this respect Case 3 was unusual in surviving for nearly six years: he was also unusual and

TABLE I  
CASES OF TRICUSPID ATRESIA WITH TRANSPOSITION OF THE GREAT VESSELS

Author	Sex	Age
Robertson (1911) .. .. .	—	9 weeks
Wason (1934) .. .. .	Male	3 months
Dunsky (1947) .. .. .	Male	5 days*
Dickson and Jones (1948) .. .. .	Male	3 months*
Robinson and Howard (1948) .. .. .	Male	6½ months
Edwards and Burchell (1949) .. .. .	Male	4 months
Kroop (1951) .. .. .	Female	5 months*
Ross (1952) .. .. .	Male	15 days
Chiche (1952), Case 9 .. .. .	—	5 weeks*
Marder <i>et al.</i> (1953), Case 3 .. .. .	Male	—*
Astley <i>et al.</i> (1953), Case P.T. .. .. .	Male	2 years
Brown and Gerlis (1954), Slide No. 52 .. .. .	Male	3 days*
Keith <i>et al.</i> (1958), 4 cases .. .. .	—	—
Ohanessian and Rodriguez (1959), Case 1 .. .. .	Male	4 years*
Macafee and Patterson, Case 1 (A.10545) .. .. .	Female	10 days*
„ Case 2 (A.14980) .. .. .	Female	7 weeks
„ Case 3 (A.15343) .. .. .	Male	5 years 10 months

\*These patients had coarctation of the aorta.

differed from Cases 1 and 2 in that the ventricular septal defect was small, while in all but 2 of the 17 cases the defect was large (Robinson and Howard, 1948; Brown and Gerlis, 1954). The high incidence of coarctation of the aorta in this group has been noted previously (Keith *et al.*, 1958) and Case 1 is a further example of this association.

The fact that 11 cases are male, out of a total of 14 in which the sex is recorded, is in keeping with the known male preponderance in transposition of the great vessels. The presence of much fenestration of the septum primum in Case 3 is an uncommon finding. Edwards and Burchell (1949) state that “defects of the atrial septum, other than patency of the foramen ovale, are the exception rather than the rule” in tricuspid atresia.

The lesions in the pulmonary vascular tree in Case 3 are of interest. Ohanessian and Rodriguez (1959) reported pulmonary vascular changes in a boy, aged 4 years, and described briefly the histological appearances as showing “some sclerosis of the pulmonary arteries”: though no illustrations were shown, this probably represents the early vascular changes of pulmonary hypertension. In Case 3 there were atheromatous plaques in the large elastic pulmonary arteries, and the media of the muscular arteries was thickened. These latter arteries showed very little intimal proliferation but this is only to be expected at this early age (Johnson, 1958). In addition this case showed two other striking pulmonary vascular lesions, namely, a necrotizing arteritis and organization of thrombi. Both these lesions are explained by the presence of pulmonary hypertension (Old and Russell, 1950). Possibly the thrombi are a consequence of the arteritis though they have been described in severe pulmonary hypertension. This is the second patient in whom pulmonary vascular lesions have been found, and both of them had survived longer than is usual in this condition.

#### SUMMARY

Three cases of tricuspid atresia with complete transposition of the great vessels and a large pulmonary blood flow have been presented.

One of these survived for an unusually long time and developed, at a comparatively young age, the histological features of severe pulmonary hypertension.

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